Appendix 4

1: Cell. 1996 Jul 12;86(1):159-71.

Related Articles, Links

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Atm-deficient mice: a paradigm of ataxia telangiectasia.

Barlow C, Hirotsune S, Paylor R, Liyanage M, Eckhaus M, Collins F, Shiloh Y, Crawley JN, Ried T, Tagle D, Wynshaw-Boris A.

Laboratory of Genetic Disease Research, National Center for Human Genome Research, National Institutes of Health, Bethesda, Maryland 20892, USA.

A murine model of ataxia telangiectasia was created by disrupting the Atm locus via gene targeting. Mice homozygous for the disrupted Atm allele displayed growth retardation, neurologic dysfunction, male and female infertility secondary to the absence of mature gametes, defects in T lymphocyte maturation, and extreme sensitivity to gamma-irradiation. The majority of animals developed malignant thymic lymphomas between 2 and 4 months of age. Several chromosomal anomalies were detected in one of these tumors. Fibroblasts from these mice grew slowly and exhibited abnormal radiation-induced G1 checkpoint function. Atm-disrupted mice recapitulate the ataxia telangiectasia phenotype in humans, providing a mammalian model in which to study the pathophysiology of this pleiotropic disorder.

PMID: 8689683 [PubMed - indexed for MEDLINE]

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